Br Heart J 1993;**69**:268–269

Pulmonary artery dissection in a patient with idiopathic dilatation of the pulmonary artery: a rare cause of sudden cardiac death

R Andrews, P Colloby, P J B Hubner

Abstract

A 31 year old man presented with a left hilar mass. Thoracic tomography showed this mass to be the pulmonary artery, and subsequently idiopathic dilatation of the pulmonary artery was diagnosed. He remained well until 11 years later when he died suddenly. Postmortem examination confirmed idiopathic dilatation of the pulmonary artery with death due to pulmonary artery dissection and cardiac tamponade.

It seems likely that idiopathic dilatation of the pulmonary artery predisposed to fatal pulmonary artery dissection.

(*Br Heart J* 1993;**69**:268–269)

Idiopathic dilatation of the pulmonary artery is an uncommon congenital cardiac defect with what is considered to be a benign prognosis and no symptoms.

Case report

A 31 year old Ugandan Asian man was referred in 1980 to the outpatient clinic after a single episode of haemoptysis. He was a lifelong non-smoker with no relevant history of occupational exposure or other past history of note, and was otherwise symptom free. Examination of the cardiovascular system and respiratory system was normal, without added clicks or murmurs. The electrocardiogram was normal and the chest x-ray showed a left hilar mass, though the cardiothoracic ratio and lung fields were normal (fig 1). This mass was thought to be the pulmonary artery and this was confirmed by subsequent thoracic tomography. Cross sectional and M mode echocardiography were normal, though the pulmonary artery could not be visualised. Idiopathic dilatation of the pulmonary artery was diagnosed. The patient declined cardiac catheterisation for confirmation. No further investigation was performed and the patient was later lost to follow up. He remained well until April 1991 when retrosternal chest pain developed which prompted him to consult his general practitioner. Two days later he died suddenly in the shower at home.

At postmortem examination the subject weighed 82 kg. The only significant findings were in the cardiovascular system. The pericardial cavity contained a large volume of fresh blood some of which had begun to clot. The heart was generally slightly enlarged (weight 407 g, left ventricle 280 g, right ventricle 90 g, ratio LV:RV 3.1:1). The pulmonary valve was morphologically normal though its circumference was 8 cm, at the upper end of the normal range. The mitral, aortic, and tricuspid valves were normal and the atria and ventricles had no intrinsic abnormalities. Important coronary atheroma was not seen. The pulmonary trunk and the main branches proximally were much dilated (maximum diameter 6 cm). When the pul-

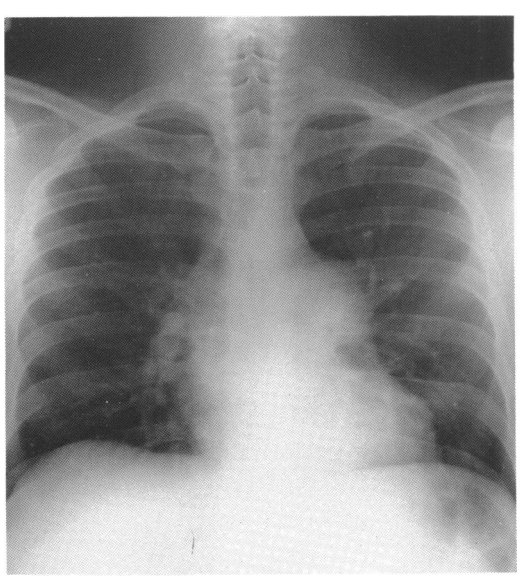

Figure 1 Chest x-ray showing a dilated pulmonary trunk.

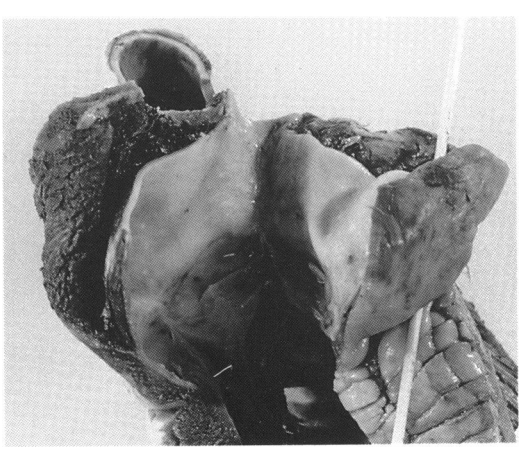

Figure 2 Macroscopic dissection of the specimen showing a greatly dilated pulmonary trunk with a dissection arising above the valve ring (probe) and extending into the pericardial cavity. The pulmonary valve and aorta are normal.

Department of Cardiology, Groby Road Hospital, Leicester
R Andrews
P J B Hubner

Department of Histopathology, Leicester Royal Infirmary
P Colloby

Correspondence to
Dr R Andrews, Department of Medicine, University Hospital, Queen's Medical Centre, Derby Road, Nottingham, NG7 2UH.

monary trunk was opened an intimal tear was seen 3 cm above the pulmonary valve ring (fig 2). A dissection extended proximally into the pericardial cavity and distally to the origin of the left and right main branches. Atheromatous plaques were seen in the pulmonary artery trunk and a large mural thrombus was present at the origin of the right main artery. Macroscopically and microscopically there was no evidence of primary lung disease or of pulmonary hypertension.

Histological examination of the pulmonary artery showed deposition of acid mucopolysaccharide in the media, similar to that seen in cystic medial necrosis but less severe. The cause of death was a dissecting aneurysm of the pulmonary trunk with subsequent haemopericardium and cardiac tamponade.

Discussion

Idiopathic dilatation of the pulmonary artery is an uncommon congenital cardiac anomaly, with one study suggesting an incidence of 6 cases per 1000 cases of congenital heart disease.[1] The condition was first described in 1923 by Wessler and Jaches,[2] though the term "idiopathic dilatation" was not coined until 1933 by Oppenheimer.[3] The diagnostic criteria suggested for idiopathic dilatation of the pulmonary artery[4 6]: (a) simple dilatation of the pulmonary trunk with or without involvement of the arterial tree; (b) exclusion of cardiac or pulmonary shunts; (c) exclusion of chronic pulmonary disease; (d) normal right heart pressures and no pulmonary valve gradient; (e) absence of arterial disease (for example, syphilis or more than minimal atherosclerosis). Common clinical features are a pulmonary ejection systolic click, a mobile widely split second heart sound, and a pulmonary regurgitant murmur in 25–80% of cases. Electrocardiographic abnormalities are uncommon and slight increases in right and left ventricular end diastolic pressure have occasionally been shown.[4] All reports conclude that the condition is benign, with the patient remaining symptom free.[7–9] The underlying pathogenesis of idiopathic dilatation of the pulmonary artery is attributed either to unequal division of the truncus arteriosus communis or to a developmental defect in the elastic tissue of the pulmonary artery.[10 11] The association of idiopathic pulmonary artery dilatation with aortic hypoplasia in rare cases supports the former theory, though the latter is more generally favoured and the description of a case of isolated peripheral pulmonary artery dilatation[12] supports it as does the finding of cystic medial necrosis in our case.

Pulmonary artery dissection is a rare complication of pulmonary hypertension.[13] Less than 40 published reports of such cases, with the most common underlying cause identified as a large left to right shunt (usually a patent ductus arteriosus), severe mitral stenosis, or primary pulmonary hypertension.[13–16] The diagnosis is almost always made at necropsy, though in two cases antemortem echocardiography showed a flap in the pulmonary artery lumen.[13 14] In one review the clinical features were found to be non-specific with 82% of patients having exertional dyspnoea, 67% retrosternal chest pain, and 52% central cyanosis. Surgical repair of pulmonary artery dissection has not been attempted. Postmortem histology often shows cystic medial necrosis in the main pulmonary artery.

1 Abbot M. Atlas of congenital cardiac disease. New York: American Heart Association, 1936.
2 Wessler H, Jaches L. Clinical roentgenology of disease of the chest. Troy, NY: Southworth, 1923:26.
3 Oppenheimer BS. Idiopathic dilatation of the pulmonary artery. *Trans Assoc Am Physicians* 1933;**48**:290–7.
4 Ramsey HW, De La Torre A, Linhart JW, Krovetz LJ, Scheibler GL, Green JR. Idiopathic dilatation of the pulmonary artery. *Am J Cardiol* 1967;**20**20:324–30.
5 Gould L, Reddy CVR, Oh KC, Kim SG. Idiopathic dilatation of the pulmonary artery. *Am Fam Physician* 1978;**17**:118–9.
6 Asayama J, Matsura T, Endo N, Watanabe T, Matsukubo H, Furukawa K, Ijichi H. Echocardiographic findings of idiopathic diltation of the pulmonary artery. *Chest* 1977; 71:671–3.
7 Liu CK, Lima AB. Enlargement of the pulmonary artery. *Med Clin North Am* 1957;**41**:119–32.
8 Deshmukh M, Guvenc S, Bentigoglio L, Goldberg H. Idiopathic dilatation of the pulmonary artery. *Circulation* 1960;**21**:710–6.
9 Balboni FA, Lopresti J. Congenital idiopathic dilatation of the pulmonary artery in children; a report of seven cases. *Bull St Francis Hosp* 1961;**17**:1–5.
10 Kaplan BM, Schlicter JG, Graham G, Miller G. Idiopathic dilatation of the pulmonary artery. *J Lab Clin Med* 1953;**41**:697–707.
11 Gold MMA. Congenital dilatation of the pulmonary arterial tree. *Arch Int Med* 1946;**78**:197–209.
12 Teitelbaum JE, Altman M. Idiopathic dilatation of peripheral pulmonary arteries. *Chest* 1978;**73**:241–2.
13 Steurer J, Jenni R, Medici TC, Vollrath TH, Hess OM, Siegenthaler W. Dissecting aneurysm of the pulmonary artery with pulmonary hypertension. *Am Rev Resp Dis* 1990;**142**:2329–21.
14 Rosenson R, St. John Sutton M. Dissecting aneurysm of the pulmonary trunk in mitral stenosis. *Am J Cardiol* 1986;**58**:1140–1.
15 Sardesai SH, Marshall RJ, Farrow R, Mourant AJ. Dissecting aneurysm of the pulmonary artery in a case of unoperated patent ductus arteriosus. *Eur Heart J* 1990;**11**:670–3.
16 Walley VM, Virmani R, Silver MD. Pulmonary arterial dissections: to be considered in patients with pulmonary arterial hypertension presenting with cardiogenic shock or sudden death. *Pathology* 1990;**22**:1–4.